Guideline for the management of Congenital Brachial Plexus Palsy

Congenital brachial Plexus injury is defined as flaccid paresis of the upper limb at birth with a larger passive than active range of motion. It is generally due to trauma to the brachial plexus at birth resulting in stretching, rupture or avulsion of some or all the nerves roots (C5-T1). Intrauterine malposition and torticollis may be a cause in some cases. Reported incidence is 1.51 to 3.2/1000 live births. CBPP is readily apparent at or shortly after birth. It is important to recognise and manage it appropriately due to the risk of long term neuromuscular dysfunction and deformities in the affected arm in some cases.

Risk factors:
shoulder dystocia, cephalic presentation of large baby (>4kg), breech delivery, prolonged second stage of labour, forceps or vacuum extraction, previous child with CBPP. One third of the cases of CBPP occur in the absence of shoulder dystocia.

Associated injuries:
Fracture clavicle/humerus, sternomastoid injury/contraction, facial/phrenic nerve palsy, transient hypoglossal, recurrent laryngeal nerve injuries

Classification: (Narakas- based on clinical findings in the first weeks of life (2 to 3 weeks)

<table>
<thead>
<tr>
<th>Narakas group</th>
<th>Other name</th>
<th>Roots involved</th>
<th>Incidence</th>
<th>Clinical features</th>
<th>Complete recovery</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Upper Erb’s palsy</td>
<td>C5, 6</td>
<td>73%</td>
<td>shoulder adducted and internally rotated, elbow extended, forearm pronated</td>
<td>3 months – 59% 6 months- 65%</td>
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<tr>
<td>II</td>
<td>Extended Erb’s palsy</td>
<td>C 5, 6, 7</td>
<td>25%</td>
<td>As above+ wrist flexed/fingers extended in ‘waiter’s tip’ position</td>
<td></td>
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<tr>
<td>III</td>
<td>Total palsy</td>
<td>C5, 6, 7, 8, T1</td>
<td>&lt;2%</td>
<td>Complete flaccid paralysis of entire upper limb- ‘flail arm’</td>
<td>3 months -0% 6 months – 14%</td>
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<tr>
<td>IV</td>
<td>Total palsy with Horner’s syndrome</td>
<td>C 5, 6, 7, 8, T1</td>
<td>25%</td>
<td>As in Narakas III + Horner’s syndrome</td>
<td></td>
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<tr>
<td></td>
<td>Klumpke’s Palsy</td>
<td>C8,T1</td>
<td></td>
<td>forearm supinated, wrist and fingers hyperextended- ‘claw hand’. Good elbow and shoulder function</td>
<td>&lt; 50%</td>
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Differential diagnosis: pseudo paralysis (fracture clavicle/humerus), epiphyseal separation of the humeral head, spinal cord injury, cervical cord lesions, congenital varicella, arthrogryposis

Evaluation:
- The initial evaluation of a neonate with suspected CBPP should include a thorough history and physical examination with emphasis also on signs of concurrent injuries and consideration of differential diagnoses.
- If CBPP is suspected, gentle handling is important. Keep head in the midline, observe resting posture, spontaneous movement, check tone, active and passive movements, reflexes (Moro, palmar grasp, tonic neck reflex) and compare both sides for symmetry. It is usually unilateral, but can be bilateral.
- Assess for Horner’s syndrome.
- Examine clavicle, shoulder, arm and neck for fracture clavicle, humerus and sternomastoid injury and torticollis.
- Assess for associated facial or phrenic nerve palsies and transient injury to recurrent laryngeal nerve (cry) and hypoglossal nerve (suck).
Management:
- Chest X ray including clavicles and humerus
- Urgent physiotherapy referral once diagnosis is established. Continued physiotherapy review and input following discharge is important. Main aim is to prevent contractures and encourage movement and strength.
- Regular outpatients follow up. Aim is to identify cases which are not likely to recover spontaneously and have residual deficits and make prompt and timely referral to specialist/multidisciplinary team for assessment, specialised investigations (electrophysiology, MRI) and surgical management as deemed necessary.
- The basic indicator for referral to a specialist or multidisciplinary team is if there is no clinical recovery of biceps function by three months of age and earlier or in cases of complete palsy and Horner's syndrome.
- Specialist/multidisciplinary referral should be made following discussion and/or review by the on-call Paediatric Neurology Consultant. Infants with total palsy and Horner's syndrome (Narakas group III and IV) should be referred urgently to the neurologist. Cases that are not improving should be discussed (but not necessarily reviewed) with the paediatric neurology consultant prior to specialist referral.
- Surgery: Primary exploration and reconstructive surgery. The optimal timing of surgical exploration of brachial plexus remains controversial; however, most agree that if it is to be undertaken, it should be done within first 6 months of life. Secondary soft tissue and bony reconstruction surgery may be required for residual deformities and deficits.

Outcome and Recovery:
Only about two-thirds of infants with CBPP have complete spontaneous recovery by 6 months. The recovery rate is better for Narakas group I and II compared to III and IV (refer the table above). Key indicator of recovery is return of biceps function. Residual deficits can occur in 20 to 33% cases. Predictors of poor outcomes are lack of elbow flexion at 3 months, complete injury and presence of Horner's syndrome.

Complications of CBPP in both untreated and treated patients include internal rotation contractures, hypoplasia of the arm, pseudo winging of the scapula, altered sensibility, muscular weakness, flexion contractures of the elbow, dislocation and importantly psychological and social consequences.

Advice to parents:
- The affected limb should be well supported with the hand, wrist, elbow and shoulder in the neutral position.
- Move the affected arm gently for washing, dressing and skin care, dress the affected arm first.
- During handling, feeding and cuddling the affected arm should be well supported.
- Parents should be fully informed about CBPP including the potential for an adverse outcome. Importance of regular follow up and physiotherapy should be emphasised.
- Information about Brachial Plexus Palsy support group: ERB’S Palsy Group, Coventry Website: www.erbspalsygroup.co.uk

References:
1 Anderson J et al. Perinatal brachial plexus palsy. Paediatr Child Health 2006; 11(2):93-100
5 Management of Obstetric Brachial Plexus Palsy. Evidence Based Practice in Paediatrics. Association of Paediatric Chartered Physiotherapists, 2001 April

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**Congenital Brachial Plexus Injury (CBPP) management flow chart**

1. **+ve Risk factors, suspected CBPP**
   - Registrar review, confirm diagnosis of CBPP
   - Documentation, CXR including clavicles and humerus (Orthopaedic referral if fracture)
   - Urgent physiotherapy referral and follow up
   - Inform neonatal consultant and GP

2. **Classify**
   - Total Palsy with Horner’s syndrome
     - **Yes**
       - Refer to Paediatric Neurologist urgently
       - Continue physiotherapy FU
     - **No**
       - Continue physiotherapy FU

3. **At 2 months FU, assess for improving biceps + deltoid function**
   - **Yes**
     - Continue physiotherapy
     - Neonatal OP FU at 3 months
   - **No**
     - Discuss with Paeds neurology
     - Continue physiotherapy

4. **Active elbow flexion**
   - **Yes, incomplete recovery**
     - Continue physiotherapy FU
     - 3 to 6 monthly at least until 12 months
     - Refer for secondary surgery, occupational therapy where appropriate
   - **Yes, complete recovery**
     - Discuss with Paeds neurology
     - Continue physiotherapy
   - **No**
     - Discharge